CASE REPORT
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Rhabdomyosarcoma of the Urinary Bladder in a Young Adult: A Case Report and Current Literature Analysis

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Abstract

Rhabdomyosarcoma (RMS) of the adult urinary bladder is an exceedingly rare malignancy, with few documented cases. While it is more common in children, its occurrence in adults is unusual and often associated with a poor prognosis due to its aggressive behavior. This report presents a 30-year-old male with hematuria. Symptoms of adult bladder RMS typically include hematuria, dysuria, and other lower urinary tract symptoms. Treatment generally involves surgery, chemotherapy, and radiotherapy, although no standardized protocol exists. This case adds to the growing body of knowledge on adult bladder RMS, highlighting the need for personalized treatment approaches.

Keywords: Bladder, hematuria, rhabdomyosarcoma, radical cystectomy

Introduction

Rhabdomyosarcoma (RMS) is a rare malignant tumor that arises from skeletal muscle progenitors and is most commonly found in children. Approximately 20% of infantile RMS cases involve the genitourinary region (1). These tumors may manifest within the bladder, prostate, paratesticular region, vagina, or uterus. In the genitourinary region, the most common manifestation of RMS is in the bladder, which often arises in the trigone area (2). The natural course of RMS typically involves rapid tumor growth with a high likelihood of recurrence, especially in adults (2). However, occurrences of RMS in the genitourinary tract among adults are exceedingly rare, with only 35 cases documented in the literature (3). Bladder RMS in adults typically manifests with symptoms such as hematuria, dysuria, or lower urinary tract symptoms (LUTS). The tumor is aggressive, with a significantly poorer prognosis in adults compared to pediatric cases (2). The optimal management often involves a combination of surgery, chemotherapy, and radiotherapy, although no standardized protocol exists for adults. This report presents the case of a 30-year-old male with bladder RMS, contributing to the limited literature on the adult population and highlighting the complexities of treatment and prognosis.

Case Presentation

A 30-year-old male presented with gross, painless hematuria and amorphous clots persisting for three months, in five episodes lasting 5–6 days each. He had a history of smoking (20 pack-years) but had quit three months prior. Physical examination and laboratory findings were unremarkable. Urine cytology indicated a low-grade urothelial neoplasm.

MRI of the lower abdomen revealed a lobulated, intraluminal mass (58x45 mm) arising from the left lateral wall of the urinary bladder. The mass was hypointense on T2-weighted images with hyperintense cystic areas, confined to the bladder wall without perivesical extension or pelvic lymphadenopathy (Figure 1).

The patient underwent transurethral resection of the bladder tumor, with histology confirming embryonal RMS. Immunohistochemistry showed positive staining for Desmin, Myogenin, and MyoD1. Fluorodeoxyglucose (FDG) positron emission tomography-computed tomography (PET-CT) scan

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showed a soft tissue lesion (7.1x5.6 cm) arising from the left lateral wall of the urinary bladder with no abnormal hypermetabolic foci elsewhere. After comprehensive evaluation, the patient underwent radical cystectomy with ileal conduit formation and bilateral pelvic lymph node dissection. The procedure and postoperative recovery were uneventful, and the patient was discharged on postoperative day seven with a follow-up plan.

Histopathology confirmed embryonal RMS of the botryoid subtype. The tumor exhibited hypercellular zones forming a cambium layer, with necrosis and atypical mitotic figures (Figure 2A, 2B). All surgical margins and regional lymph nodes were tumor-free. Immunohistochemistry reaffirmed Desmin and Myogenin positivity (Figure 2C, 2D).

Verbal and written informed consent was obtained from the patient for the case report.

Discussion

RMS is a rare and aggressive malignancy originating from mesenchymal tissues, typically affecting children with a slight male predominance (4). However, its occurrence in adults,

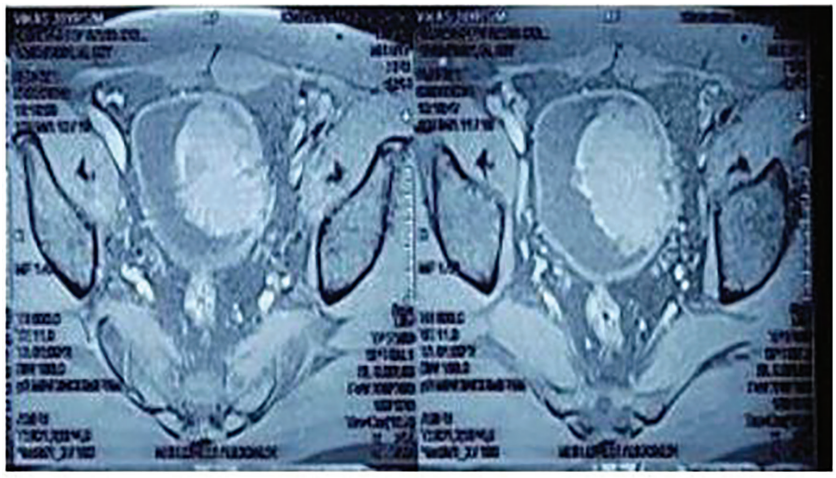

Figure 1. MRI of the lower abdomen revealed an intraluminal, lobulated mass originating from the left lateral wall of urinary bladder
MRI: Magnetic resonance imaging

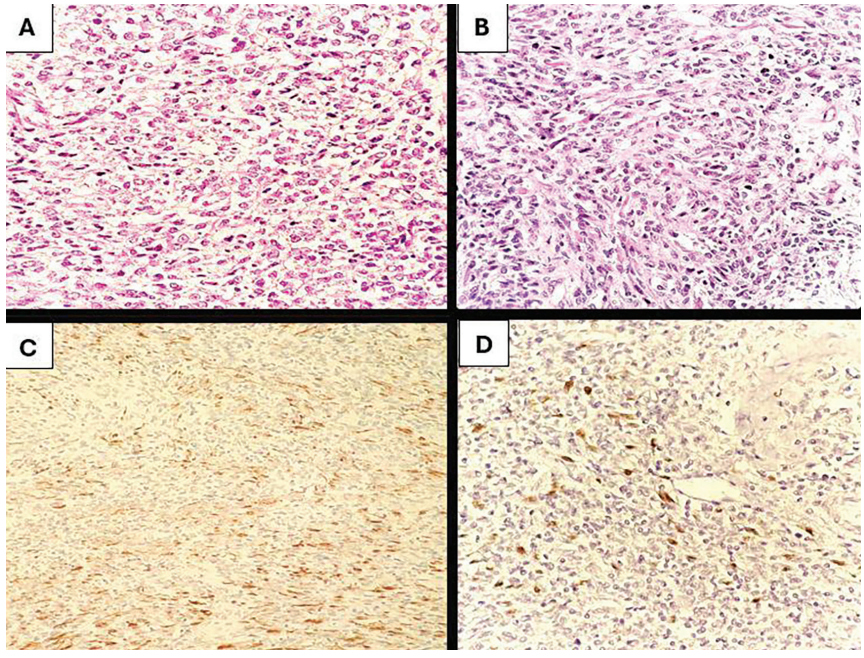

Figure 2. Hematoxylin and eosin stain demonstrating tumor cells (A) and spindle or round cells with Rhabdomyoblastic differentiation (B). Immunohistochemistry showing positive staining for Desmin (C) and Myogenin

particularly in the urinary bladder, is exceedingly uncommon, contributing to diagnostic challenges and treatment uncertainties. Our case reviews a young male diagnosed with bladder RMS.

Bladder RMS is rare and has been reported in only a small number of adult cases. The literature indicates that RMS in the bladder accounts for less than 2% of all urogenital sarcomas in adults (5). Leiomyosarcomas make up about 50% of sarcoma cases, while RMS represent roughly 20% (2,5).

Bladder RMS often presents with nonspecific urinary symptoms, such as hematuria, voiding LUTS, or urinary retention as observed in most cases (6). Dysuria, constipation, and pelvic pain typically develop at a later stage (2,7). In our case, the patient presented with gross hematuria, which is the most common initial symptom reported in adult bladder RMS.

Histologically, RMS is classified into different subtypes: embryonal, alveolar, and pleomorphic. The botryoid variant of embryonal RMS, the subtype diagnosed in our case, is the most common and is characterized by a better prognosis compared to alveolar or pleomorphic variants (8,9). Immunohistochemically, RMS typically exhibits positive staining for Desmin, along with MyoD1 and/or Myogenin (10). Consistent with this, our study found Desmin and Myogenin to be positive.

Adult RMS is characterized by small, round blue cells along with large, random anaplastic cells. In some cases, signs of striated muscle differentiation are observed, with cells exhibiting a more abundant, eosinophilic cytoplasm. These cells are referred to as "tadpoles" or "tennis-racket" cells (11,12).

Treatment for adult bladder RMS lacks standardization due to its rarity. While multimodal approaches (surgery, chemotherapy, radiotherapy) are effective in children, they are less successful in adults (3). Given the localized stage of RMS in our patient, we decided to proceed with radical cystectomy followed by adjuvant chemotherapy. Complete surgical resection, when feasible, provides the best chance for disease control and long-term survival (3,13).

Chemotherapy for RMS typically involves regimens like Vincristine, Actinomycin-D, Cyclophosphamide or Vincristine, Actinomycin-D, Ifosfamide based on risk, achieving high response rates in localized cases, particularly in embryonal RMS. These regimens improve survival in low to intermediate-risk cases (11).

The patient was referred to another center and scheduled for 12 cycles of adjuvant chemotherapy using the VDC regimen (Vincristine, Doxorubicin, and Cyclophosphamide). At the time of this report's submission, the patient had completed 8 cycles. A follow-up FDG PET-CT scan was performed after 6 cycles which revealed no abnormal FDG avidity, indicating no signs of systemic recurrence. Ongoing follow-up care, including additional cycles of chemotherapy, will be critical in monitoring for any potential recurrence, particularly given the aggressive nature of RMS and the challenges posed by managing such rare cases in adults.

A systematic review done on adult bladder RMS emphasizes the high recurrence rate (36.4%) among patients, despite various treatment strategies; and suggests that radical cystectomy may not offer a significant survival advantage over partial cystectomy, supporting the potential for bladder preservation in select cases. Furthermore, this review underscores the importance of multimodal therapies, such as chemotherapy and radiotherapy, particularly when managing high-risk tumors . Our case contributes to this broader discussion by detailing a radical cystectomy approach in a patient with embryonal RMS, which aligns with the treatment regimens discussed in the review (14).

The prognosis for adult bladder RMS is poor. The 5-year survival rates of adult RMS are 17% for kidney, 22% for bladder, and 33% for prostate. In contrast, pediatric bladder RMS has a 66% survival rate (3,13). Regardless of stage or treatment, adult RMS typically results in survival of 3 to 19 months post-treatment (15).

Conclusion

Bladder RMS in adults is rare and challenging, with cystectomy being the standard treatment for localized cases. The role of chemotherapy and radiotherapy for metastatic disease is still unclear. Our case adds to the literature, showing that radical surgery can lead to successful outcomes.

Ethics

Informed Consent: Verbal and written informed consent was obtained from the patient for the case report.

Footnotes

Authorship Contributions

Surgical and Medical Practices: A.S., U.S., S.A.S., Concept: U.S., A.M., Design: A.S., N.M.P., S.A.S., Data Collection or Processing: A.S., N.M.P., S.A.S., Analysis or Interpretation: A.S., U.S., A.M., Literature Search: A.S., U.S., A.M., Writing: A.S., N.M.P., S.A.S.

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